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## Idiopathic pulmonary artery aneurysm and its surgical management: A rare case report

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### Abstract

Pulmonary artery aneurysms are rare and infrequently diagnosed. Various origins have been described, like congenital causes, acquired causes, and idiopathic. Among all of them idiopathic pulmonary artery aneurysm is the rarest variety.

Clinical manifestations remain nonspecific; include dyspnoea, chest pain, hoarseness, palpitation. If bronchial compression is there, it may cause cough, breathlessness, cyanosis, syncope attacks and rarely large aneurysm can cause haemoptysis.

There is no clear guideline for the treatment for pulmonary artery aneurysm. Conservative treatment is the main stay for this disease when it is not related with any congenital anomalies and normal pulmonary artery pressure. In case of disabling symptoms, operative management is the cornerstone for this entity.

**Keywords:** pulmonary artery, angiography, aneurysm, idiopathic

### Introduction

Pulmonary artery aneurysm (PAA) is defined as dilation of the proximal pulmonary artery or one of its branches. It is difficult to diagnose and often is an autopsy finding. It is classified as central, which arises from the main pulmonary artery, or peripheral, which arises from segmental or sub-segmental arteries. In computed tomography, the upper limit for adults of the pulmonary artery trunk diameter is 29 mm, and the upper limit of the branch pulmonary artery is 17 mm. The pulmonary hypertension is an important risk factor for development and rupture of PAA.

Idiopathic PAA formation is rare, but an increasing number of cases are being reported in the literature [1-3] Greene and Baldwin<sup>4</sup> have defined 4 pathological criteria for an idiopathic PAA: simple dilatation of the pulmonary trunk with or without involvement of the rest of the arterial tree, the absence of intracardiac or extracardiac shunts, the absence of chronic cardiac or pulmonary disease, and the absence of arterial disease such as syphilis or more than minimal atheromatosis or arteriosclerosis of the pulmonary vascular tree.

Conservative management is appropriate for asymptomatic cases and without underlying cardiac pathology but surgical treatment is the main stream for pulmonary artery aneurysm. The most common procedure is the replacement of the Pulmonary artery and the pulmonary trunk with synthetic graft (Gore-Tex or Dacron tube), homograft's, or engrafts (porcine aortic grafts or bovine jugular conduits) We did surgical excision of pulmonary artery aneurysm and reconstruction of native vessel without grafts, preserving valvular function.

### Case report

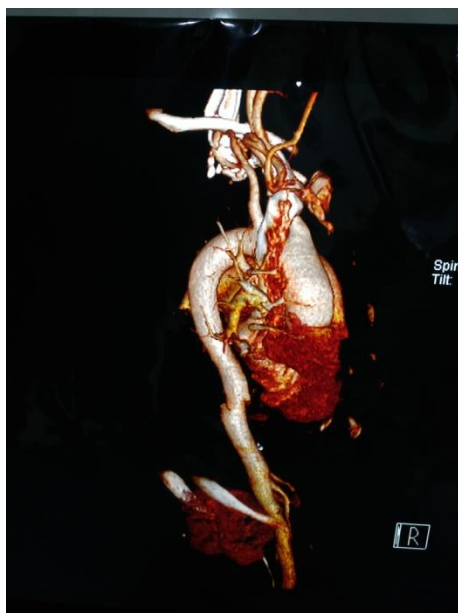
A 50 year old female patient, presented with complains of having chest pain and dyspnoea for 1 year. Initially it presented on exertion which gradually increased in its intensity, and became evident even at rest for last 20 days. Patient had no history of any other complains or co-morbidities. Patient is not on any medications or no positive cardiovascular anomaly.

On examination patient was having bilateral equal air entry sound and having normal cardiac sounds and rhythm. Chest x ray (Figure 1) shows well defined radio-opacity at left peripheral region. 2D echo showed pulmonary artery aneurysm with near normal pulmonary valve except leaflet thickening. She had mild PS, PR without clot or vegetations and 60% LVEF. HRCT thorax shows markedly dilatation of main pulmonary trunk and left pulmonary artery suggestive of aneurysm. Findings were confirmed with pulmonary CT angiography (Figure 2). Maximum

diameter of main pulmonary trunk was 4.2 cm, left pulmonary artery was 5.8 cm and right pulmonary artery was 2.5 cm. CAG report showed normal coronary and LV functions.



**Fig 1:** Pre-op chest x ray PA view



**Fig 2:** Pulmonary CT angiogram

### Surgical technique

Mid line sternotomy was done. #22 aortic cannula and double stage atrial venous cannula inserted after adequate heparinization. Patient cooled to 30 degree centigrade. Ante grade root cardiopligia given after aortic cross clamp to attain cardiac arrest. Main pulmonary artery (MPA), right and left pulmonary artery exposed and dissected (Figure 3). Right and left pulmonary looped. The MPA and LPA opened on stay sutures. No thrombus found. Pulmonary valve thickened but neither stenosis nor regurgitant. Pulmonary valve Annulus was normal.

Excess Pulmonary artery wall excised after sizing with 22 hegars dilator. MPA and LPA are reconstructed by primary suturing of edges using 4-0 prolene with tanned pericardial patch reinforcement. Complete hemostasis maintained (Figure 4).

Cross clamp removed after deairing. Cardiac rhythm reverted in sinus. The Cardiac decanulation done in stages. Heparin

reversed using adequate quantity of protamine. One ventricular pacing wire placed. Mediastinal drainage tube inserted. Sternum closed using #6 three SS wire .skin and subcutaneous tissue sutured using monocryl and vicryle respectively. Post op period uneventful. (Figure 5 and Figure 6).



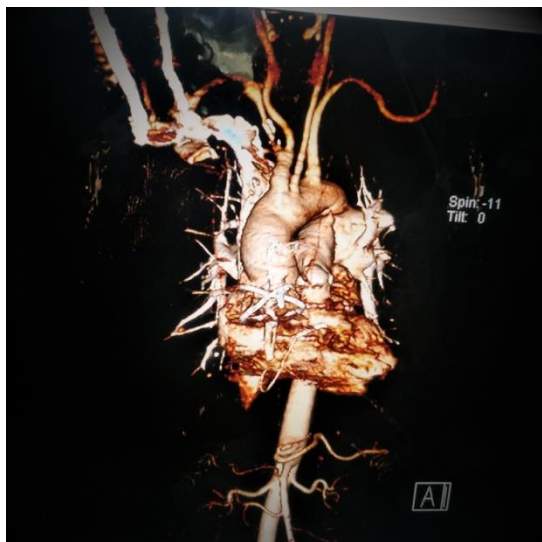
**Fig 3:** Pulmonary artery aneurysm



**Fig 4:** Pulmonary artery after repair



**Fig 5:** Post op chest xray



**Fig 6:** Post op pulmonary CT angiography

### Discussions

Aneurysms and pseudo aneurysms are rare abnormalities of the pulmonary arteries. Pulmonary Artery Aneurysm (PAA) is a rare condition with an incidence of 1 in 14,000 autopsies [3]. Plotnick, define a PAA as a PA with a diameter exceeding 40 mm, distinguishing between an ectasia of the PA and a true PAA. Unlike aortic aneurysm it has equal prevalence in both genders and occurs in younger patients [5]. The likely pathology seems to be increased hemodynamic stresses combined with underlying vessel wall weakness [5-6]. It is commonly associated with congenital heart defects (shunts, PDA or VSD), pulmonary artery hypertension and connective tissue disorders such as Marfan's syndrome and infections like syphilis and Tuberculosis [6]. It typically presents with haemoptysis, chest pain, cough and/or dyspnoea but is also not infrequently discovered as an incidental finding [7]. The advents of contrast-enhanced CT, pulmonary angiography and echocardiograms have assumed a primary role in diagnosis [7]. The complications may include pulmonary hypertension, thrombus formation, bronchial compression or aneurysmal rupture. Management is surgical coupled with treatment of the underlying disorders.

Our case was rarest variety among all PAAs. As it was idiopathic PPA fulfilling all 4 criteria<sup>4</sup> i.e simple dilatation of the pulmonary trunk without involvement of the rest of the arterial tree, without intracardiac or extracardiac shunts, chronic cardiac or pulmonary disease, and in the absence of arterial disease such as syphilis or more than minimal atheromatosis or arteriosclerosis of the pulmonary vasculature.

Patient was having dyspnoea at rest and chest pain at presentation, pulmonary CT angiography shows max diameter 4.2 cm of main pulmonary trunk and 5.2 cm of left pulmonary trunk suggestive of pulmonary artery aneurysm.

As our patient was symptomatic, and investigations suggestive of significant enlarged pulmonary artery aneurysm, decided to go for surgical management. The most common procedure is the replacement of the PA and the pulmonary trunk with a conduit starting in the right ventricular outflow tract. This can be performed with Gore-Tex or Dacron tubes, homografts, or xenografts (porcine aortic grafts or bovine jugular conduits). But here we took novel approach. We reconstructed PA by primary suturing of edges, after cutting excessive portion of wall, using 4-0 prolene with tanned pericardial patch reinforcement, to avoid graft related complications.

### Conclusion

Idiopathic PAAs seldom occurs, are rarely diagnosed, and do not present with distinct symptoms. To date, there are no clear guidelines or rules on the optimal treatment for patients with PAAs because of the small number of cases.

Conservative treatment is appropriate for asymptomatic patient, having normal PA pressure and pulmonary artery diameter less than 3.5 cm. Such patients should be re-evaluated regularly, and a change in treatment should strongly be considered in case of compression of adjacent structures, thrombus formation in the aneurysm sack,  $\geq 5$ -mm increase in the diameter of the aneurysm at 6 months.

The appearance of clinical symptoms, evidence of valvular pathologies or shunt flow, or verification of PAH strongly indicates surgical management because of the possible fatal outcome most common being rupture of aneurysm, compression of the surrounding structures.

### Declarations

Funding: none

Conflict of interest: none

Ethical approval: taken

### Abbreviations

PA- pulmonary artery

PAA-Pulmonary artery aneurysm

LVEF-left ventricular ejection fraction

PAH-pulmonary artery aneurysm

LV- left ventricle

CAG-Coronary angiography

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